Young Male with Syncope, Absent Lower Limb Pulses, Hypertension and Systolic Murmur

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Fig. 1: Plain frontal chest radiograph- minimal cardiomegaly with notching and erosion of posterior lower rib margins (arrows).

Fig. 2: CT Aortogram showing non-opacification (arrow) of aorta just distal to origin of a dilated left subclavian artery suggestive of coarctation.

Fig. 3: Dilated tortuous internal mammary and intercostal arteries on CT aortogram.

Fig. 4: Aorta distal to coarctation showing good contrast opacification, filling through multiple collaterals (arrow).

An otherwise healthy 18 year old male presented with one episode of syncope the previous night, with no associated symptoms. Examination revealed supine brachial blood pressure (BP) of 180/110 mm of Hg in both arms without significant postural drop. Bilateral upper limb pulses were normal in rate, rhythm, volume without any radiodermal delay. However, all lower limb pulses were bilaterally absent. A grade 2 high pitched ejection systolic murmur was auscultable in the left third intercostal space. Rest of the examination (including abnormal arterial pulsations or bruits) was unremarkable. A 12-lead electrocardiogram revealed left axis deviation with left anterior hemiblock. Chest x-ray showed minimal cardiomegaly with rib notching (Fig. 1). Echocardiography demonstrated coarctation of aorta (CoA) with discrete narrowing noted immediately after origin of left subclavian artery (LSCA) with pressure gradient of 60 mm of Hg. A bicuspid aortic valve without left ventricular outflow tract gradient was also noted. CT Aortogram revealed non-opacification of 4 mm segment of aorta distal to LSCA suggestive of CoA (Fig. 2). Bilateral subclavian and internal mammary arteries were dilated, tortuous (Fig. 3) with multiple chest and abdominal wall collaterals (Fig. 4); both renal arteries were normal. The patient was posted for surgical coarctoplasty after BP control with multiple anti-hypertensives.

CoA is the most common congenital cardiovascular cause of hypertension with incidence 1–8 per 1000 live births. The most common associated congenital malformation is a bicuspid aortic valve that imposes inherent risks. The mean survival of untreated patients is 35 years. Intervention may be surgical (gold standard) or percutaneous. The 20-year survival rate is 91% if coarctation is repaired before 14 years of age, and 79% if repaired after that age. Even when the anatomic lesion is corrected, up to 30% of patients develop subsequent/persistent hypertension and are at risk of accelerated coronary artery disease and cerebrovascular events.

Reference

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